Original Article

Anomalous Origin of One Pulmonary Artery from the Ascending Aorta. Surgical Repair Resolving Pulmonary Arterial Hypertension

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Objective

To emphasize the diagnostic possibility of the anomalous origin of one pulmonary artery from the ascending aorta in infants with clinically refractory heart failure and no intracardiac structural defect.

Methods

Retrospective study of 4 infants with refractory heart failure undergoing 2-dimensional echocardiographic study with subcostal, suprasternal, and parasternal views, and hemodynamic and angiocardiographic study in the anteroposterior projection.

Results

Three of the 4 infants had their right pulmonary artery originating from the ascending aorta as their major diagnosis. In the fourth patient, the left pulmonary artery originated from the ascending aorta in association with a large interventricular septal defect. The pressure level in both pulmonary arteries in all infants was that of the systemic level. All patients underwent surgery, which consisted of translocation of the anomalous pulmonary artery from the aorta. Neither immediate nor late cardiac deaths occurred.

Conclusion

Once the diagnosis of anomalous origin of the pulmonary artery from the ascending aorta in the isolated form is established, the surgical correction should be immediately performed, not only because of the risk of developing pulmonary vascular disease, but also because of the excellent surgical results currently obtained.

Key words

anomalous pulmonary artery, obstructive pulmonary vascular disease, acyanotic congenital heart disease, increased systemic pulmonary flow

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Pulmonary artery originating from the ascending aorta is a rare, frequently fatal malformation if early surgical repair is not performed. In 1868, Fraentzel [1] reported a case for the first time. The origin of the right, and less frequently of the left, pulmonary artery from the aorta occurs in the presence of separated aortic and pulmonary valves, and should be differentiated from the truncus arteriosus, in which the pulmonary arteries originate from the ascending aorta, but in the presence of a common semilunar valve.

The origin of the pulmonary artery from the ascending aorta is responsible for a great left-to-right shunt at the systemic-pulmonary level.

The contralateral lung receives all the cardiac output in addition to the occasional blood flow of the associated anomalies, such as ductus arteriosus, aortopulmonary window, and interatrial and interventricular septal defects, which may occur in approximately 40% of cases [2-4] (tab. I). The clinical manifestation usually occurs in the infant, or, more rarely, in the newborn as respiratory distress or congestive heart failure due to increased pulmonary resistance [5-10]. The diagnosis may be established by use of 2-dimensional Doppler echocardiography [11,12]. Additional information is provided with cardiac catheterization and cineangiocardiography [5]. Without surgical correction in the first year of life, pulmonary arterial hypertension may develop in both lungs independently of the right or left anomalous origin of the pulmonary artery.

Methods

A retrospective study of 4 infants with anomalous origin of the pulmonary artery from the ascending aorta was carried out. Their age ranged from 2 to 6 months, and 3 were of the male sex. On presentation, all infants were acyanotic in clinically refractory heart failure. All underwent 2-dimensional echocardiography and color-flow Doppler. The most frequently used views were suprasternal, subcostal long-axis, and transverse parasternal. The pressures in the pulmonary arteries and aorta were determined in the hemodynamic study. On angiography, contrast medium was injected into the right and left ventricles and into the aorta in the anteroposterior view. The resistances in the pulmonary arteries and aorta could not be determined due to technical problems.

Results

Of the 4 patients, 3 were diagnosed with anomalous origin of the right pulmonary artery from the ascending aorta by use of only

Table I - Anomalous origin of one pulmonary artery from the ascending aorta in 197 cases reported in the literature

	n	Age	AORPA(n)	AOLPA(n)	PDA (%)	IVD (%)	IAD/FO (%)	AAo (%)	TF (%)	Mortality	Follow-up
Fontana [2]	65	-	65	0	68%	8%	16%	11%	3%	-	-
Kutsche et al [3]	108	-	83	16	57%	0%	9%	18%	16%	-	-
Nouri et al [4]	25	124 d	21	3	56%	0%	48%	20%	12%	3.4%	23

AORPA - anomalous origin of the right pulmonary artery; AOLPA - anomalous origin of the left pulmonary artery; PDA - patent ductus arteriosus; IVD - interventricular septal defect; IAD/FO - interatrial septal defect/patent foramen ovale; AAo - anomaly of the aortic arch; TF - tetralogy of Fallot.

echocardiography (fig. 1). The angiographic study (fig. 2) confirmed the major anomaly and diagnosed as a secondary anomaly a small interatrial septal defect in 2 patients, and a persistent ductus arteriosus in the third patient (tab. II). The diagnosis of anomalous origin of the left pulmonary artery from the ascending aorta was established on echocardiography (fig. 3) and confirmed on cineangiocardiography (fig. 4). This latter patient had a wide interventricular septal defect as a secondary anomaly. Blood pressure assessment in the 4 patients showed systemic levels in the anomalous pulmonary artery, as well as in the contralateral pulmonary artery (tab. III).

Stenoses at the origin or trajectory of the anomalous pulmonary artery, as well as at the contralateral pulmonary artery, were observed neither on angiography nor on blood pressure study of the 4 patients. All patients underwent surgical correction, performed using translocation of the anomalous pulmonary artery from the aorta.

The surgical technique used consisted of establishing extracorporeal circulation, aortic clamping, and infusion of the solution for cold crystalloid cardioplegia. Then, the anomalous pulmonary artery was sectioned close to the aorta, and a terminolateral anastomosis was established between the anomalous pulmonary artery and the pulmonary trunk. In the patient with the anomalous origin of the left pulmonary artery from the ascending aorta, the interventricular septal defect (18 mm) was repaired with a patch of bovine pericardium. Except for patient number one, who died due to an accident 2 years after surgery, no other immediate or late deaths occurred.

After translocation of the anomalous pulmonary artery, pressure in both pulmonary arteries in all patients became normal. The postoperative echocardiographic study ruled out the presence of stenoses at the site of anastomosis.

Discussion

The anomalous origin of one pulmonary artery from the ascending aorta is a defined and well-known entity [7]. It is a malfor-

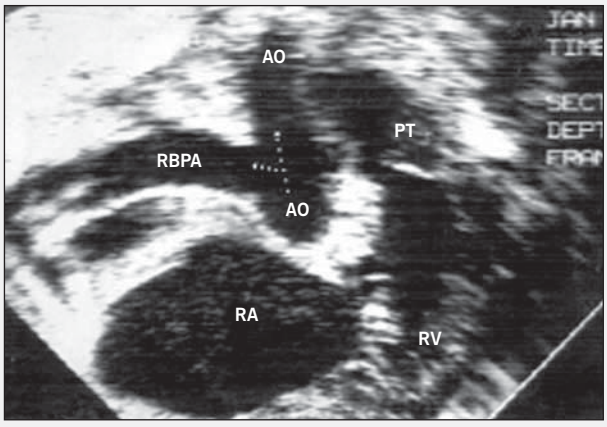

Fig. 1 - Subcostal long-axis view showing dilation of the right ventricle and of the pulmonary trunk. Visualization of the aorta allows identification of the right branch of the pulmonary artery, also dilated, originating from the proximal portion of the ascending aorta.

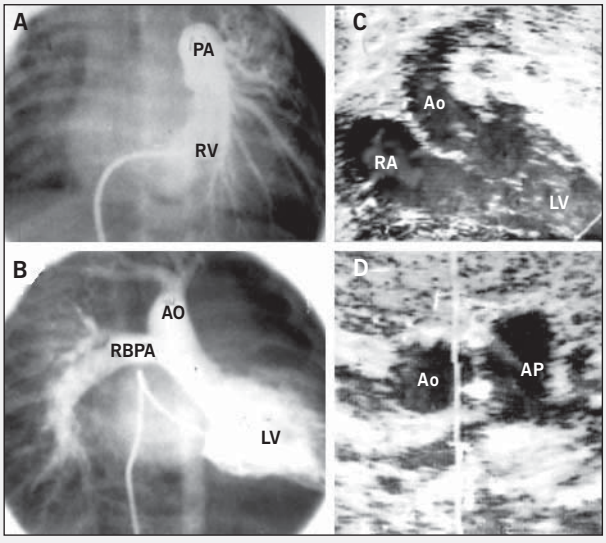

Fig. 2 - A) Right ventriculography during systole. Opacification of the very dilated pulmonary trunk and of its left branch. The right branch of the pulmonary artery cannot be seen; B) left ventriculography. The aorta emerges from this cavity, from which the also dilated right branch of the pulmonary artery originates; C) subcostal view: the abnormal origin of the right pulmonary artery from the ascending aorta cannot be seen. D) Postoperative 2-dimensional echocardiogram, transverse parasternal view: presence of normal bifurcation of the pulmonary trunk.

Table II - Major findings in 4 patients with anomalous origin of one pulmonary artery from the ascending aorta

Case	Age	Sex	Major anomaly	Secondary anomaly	Manifestation	Diagnosis	Surgical treatment	Follow-up
1	2 m	F	AORPA	IAD	CHF	ECHO/CAT	Ao-PA translocation	Death after 2 years – accident
2	6 m	M	AOLPA	IVD	CHF	ECHO/CAT	Ao-PA translocation	2 years
3	3 m	M	AORPA	-	CHF	ECHO/CAT	Ao-PA translocation	10 years
4	5 m	M	AORPA	PDA	CHF	ECHO/CAT	Ao-PA translocation	4 years

AORPA - anomalous origin of the right pulmonary artery; AOLPA - anomalous origin of the left pulmonary artery; PDA - patent ductus arteriosus; IVD - interventricular septal defect; IAD - interatrial septal defect; CHF - congestive heart failure; ECHO - echocardiography; CAT - angiographic study; Ao - aorta; PA - pulmonary artery.

mation completely different from those in which the pulmonary arteries originate from the aorta via ductus arteriosus or are supplied through the collaterals between the systemic and pulmonary arteries [13]. The morphology of the anomalous pulmonary artery is constant: it originates from the wall of the ascending aorta, 5-30 mm above the ventriculoarterial junction [14]. Kutsche and Van Mierop [3], in a multicenter study about the pathogenesis and associated anomalies in 108 cases, reported that the right pulmonary artery rarely originates from the lateral portion of the ascending aorta juxtaproximal to the right brachiocephalic trunk. The incidence of the anomalous origin of the right pulmonary artery is not only more frequent, but also is not pathogenically related to the anomalous origin of the left pulmonary artery. Yet, the anomalous origin of the right pulmonary artery close to the right brachiocephalic trunk is pathogenically different from the type originating close to the aortic valve [3]. In regard to pathogenesis, some authors believe that the anomalous origin of the left pulmonary artery from the ascending aorta is an anomaly of the aortic arch, because, in all cases studied, tetralogy of Fallot or anomalies of the aortic arch, or both, were associated. In our only case (patient number 2), the only associated malformation was interventricular septal defect. Except for the patients with tetralogy of Fallot as an associated defect, the size of the anomalous artery and of the contralateral pulmonary artery is the same.

The first successful surgical correction was performed by Armer et al [15] in 1961 using a Dacron graft placed between the right pulmonary artery and the pulmonary trunk [15]. Kirkpatrick et al [6] performed the first anatomical correction with translocation of the right pulmonary artery to the pulmonary trunk [6].

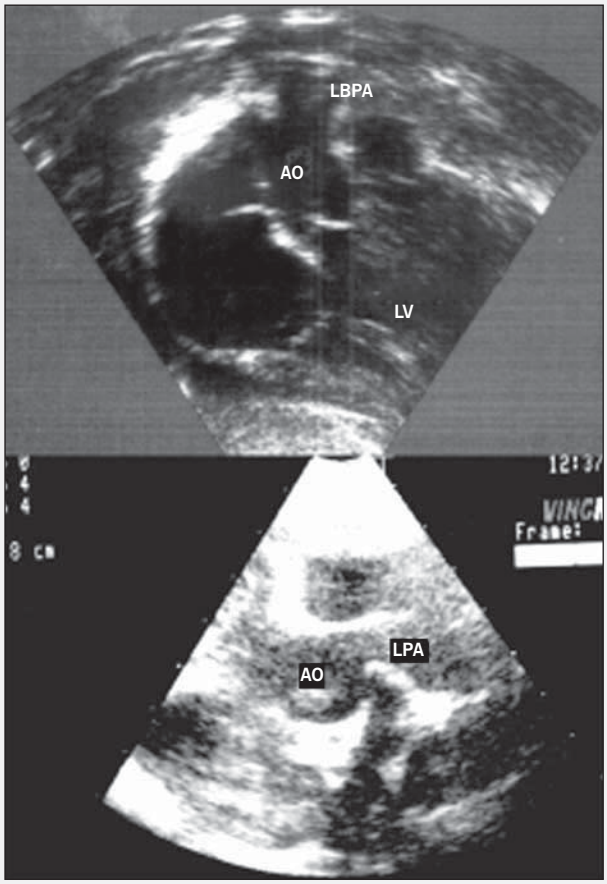

Fig. 3 - Upper) Subcostal long-axis view. The aorta originates from the left ventricle; from the proximal portion of the aorta, the dilated left branch of the pulmonary artery originates; Lower) Suprasternal view. A great-caliber vessel is clearly seen originating from the ascending aorta and running to the posterior region - left branch of the pulmonary artery.

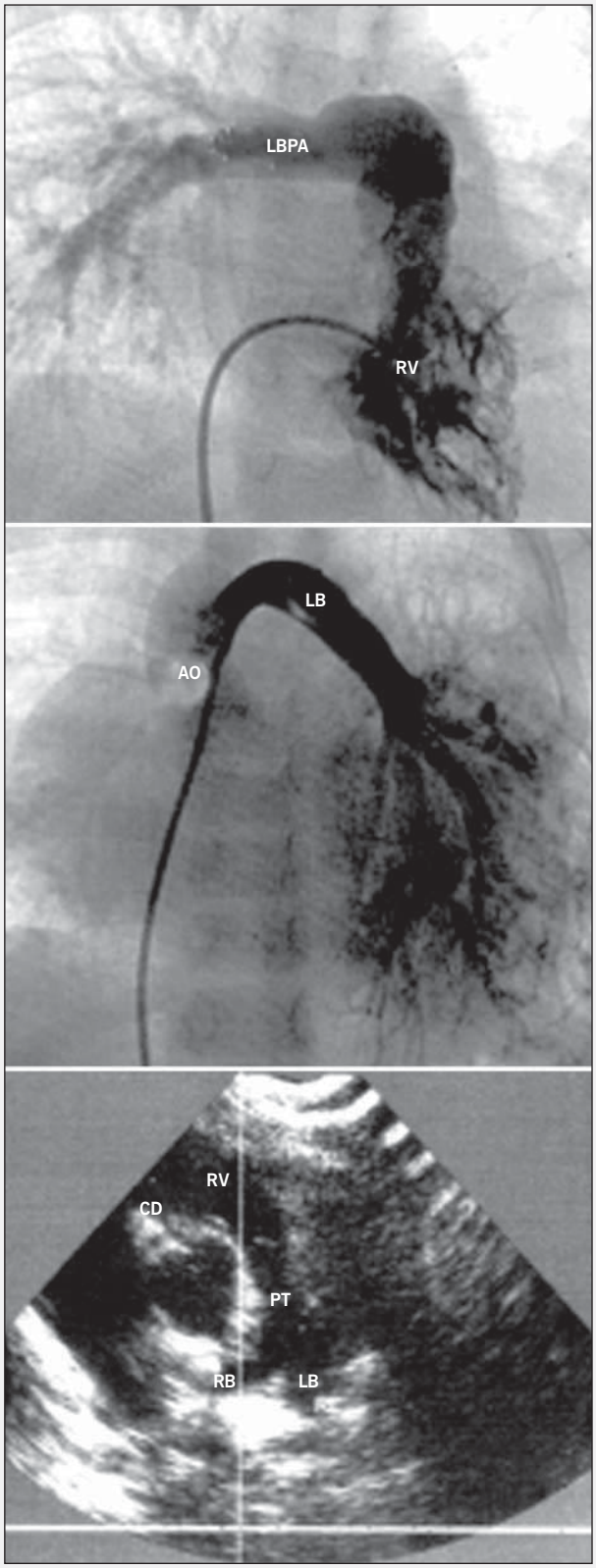

Fig. 4 - Upper: Right ventriculography showing the very dilated pulmonary trunk and right branch of the pulmonary artery. The left branch of the pulmonary artery is not seen; Central: Aortography showing the origin of the left pulmonary artery from the ascending aorta; Lower: Transverse view. The left branch of the pulmonary artery is connected to the pulmonary trunk.

Table III - Preoperative pressure levels of the 4 patients obtained on cardiac catheterization

Case	RBPA	LBPA	Ao	RV
1	90/50	100/55	105/55	100/18
2	90/40	85/40	100/60	100/16
3	95/45	90/50	95/50	100/18
4	88/50	90/45	100/55	90/14

RRBPA - right branch of the pulmonary artery; LBPA - left branch of the pulmonary artery; Ao - aorta; RV - right ventricle.

The diagnosis may be established on suprasternal views of 2-dimensional echocardiography [11,12] when a vessel originates posteriorly from the ascending aorta. In these cases, the possibilities of aortopulmonary window and truncus arteriosus should be ruled out. Thus, lack of the normal bifurcation of the pulmonary trunk is required. In situations in which the anomalous pulmonary artery originates from the lateral face of the ascending aorta, the subcostal views allow the diagnosis to be made very safely [16].

In the preoperative assessment, a careful analysis of the physiopathology of the right and left lung, and of the right ventricle is necessary. When the anomalous origin of the right or left pulmonary artery is not associated with intracardiac defects, 2 separate nonparallel pulmonary circuits with independent flow and resistance exist [17]. Considering, initially, the right lung, the right pulmonary artery will have systemic blood pressure levels and a flow that will depend on the resistance of the right pulmonary vascular system with elevated pressure. In the left lung, the pulmonary flow will be equal to that of the systemic venous return. Variations in left pulmonary vascular resistance will be transmitted to the right ventricle, and, consequently, to the total cardiac output. The reasons for the elevation in left pulmonary vascular resistance are not yet well understood; it seems not to result from an isolated increase in pulmonary flow, because in patients with congenital absence of one lung and who also have one right ventricle managing the entire cardiac output, no major elevation in pulmonary vascular resistance is observed in their single lung [18].

Except for the flow, the following factors can modify pulmonary vascular resistance: 1) pulmonary arterial anatomy, such as diameter, number and distribution of the vessels; 2) vascular tonus; and 3) blood rheology. Our patients had no evidence of abnormal blood elements that could affect the characteristics of normal flow. The anatomical obstructions were also ruled out by measuring pulmonary capillary wedge pressure.

Just for speculation, the physiopathological events in the right lung may have been produced by neurovascular reflexes or humoral vasoactive mediators, or both –excessive resistance in the left lung. These questions could justify the drop in pulmonary artery pressure (regardless of whether the anomalous pulmonary artery is right or left) immediately after surgical correction. Therefore, the immediate drop in pulmonary artery pressure favors a reflex mechanism, although a humoral factor cannot be ruled out. Despite all these considerations, the pulmonary blood flow seems to be the critical factor in the development of pulmonary vascular disease, because patients with tetralogy of Fallot as an associated defect show only unilateral alterations.

The elevation in pulmonary vascular resistance caused some difficulty in the echocardiographic diagnosis of the patient with anomalous origin of the left pulmonary artery from the ascending aorta, because the patient also had an associated wide interventricular septal defect. However, the angiocardiographic study established the diagnosis. The pulmonary biopsy did not provide further information about the vascular differentiation between the abnormally perfused and the contralateral lungs [5,19]. However, other investigators reported greater vascular changes in the contralateral lung [20,21]. These alterations were attributed to the increased totally oxygenated blood flow in the abnormally perfused lung.

Early surgical intervention, preferentially before 12 months of age, is very important to prevent the development of irreversible pulmonary vascular disease [22]. In patients in whom the anomalous origin is an isolated finding, surgical correction should be considered as soon as the diagnosis is established, preferentially within the first 6 months of life, due to the possibility of the early development of pulmonary vascular disease. The surgical correction may be performed with or without extracorporeal circulation, depending on the technical difficulties found during the procedure [13,14]. In patients with associated tetralogy of Fallot, the surgical risk is greater; however, successful surgical repair has been reported in those patients [23,24]. Hypoplasia of the pulmonary artery connected to the right ventricle can protect the lung from hypertensive vascular disease, which does not occur in the abnormally perfused lung.

In conclusion, the results of the surgical correction in the past years have been positive, except in patients with associated cardiac malformations. In our case series, no death was observed during surgery or later. The palliative treatment, such as pulmonary artery banding, and ligation of the associated ductus arteriosus and aortopulmonary shunt, increases mortality substantially.

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